Acquired cholesteatoma in children following congenital cholesteatoma surgery

Mauricio A. Cohen a,*, Maya A. Kuroiwa a, Robert G. Berkowitz b,c

a Department of Otolaryngology, Faculty of Medicine, University of Chile, Chile
b Department of Otolaryngology, Royal Children’s Hospital, Melbourne, Australia
c Department of Paediatrics, Royal Children’s Hospital, Melbourne, Australia

1. Introduction

Congenital cholesteatoma (CC) is relatively uncommon, representing approximately 4% of childhood cholesteatomas [4], but has been well described in the medical literature for over 40 years, since Derlacki and Clemis established its diagnostic criteria and contemporary understanding back in 1965 [1]. Most of the series published up to now have a short term follow up, focusing primarily upon diagnostic issues, surgical techniques for early lesions, recurrence or residual disease management and development of classification systems [2,3], and only few cohorts have a close and long follow up.

Nevertheless, it has been our experience that while many patients experience an uneventful recovery after surgery for CC, there is a significant subgroup of patients that in the long term suffer from acquired pathology of the middle ear. These long-term complications, only hinted previously [4], have been properly recognized recently in an article by Lazard et al. [5], where 16 out of 117 CC patients were reported to develop a retraction pocket (RP) after a median time of 15 months post-surgery. However, this represented only a 13.7% of all patients, even when including patients that presented initially with otorrhea, which as they suggest, can represent acquired disease from the beginning, furthermore, their main attention was on risk factors, and therefore omitted any mention of the significance these RP represented for the patients or the management utilized afterwards.

The purposes of our report are to present acquired middle ear disease, including cholesteatoma, can follow surgical removal of CC, and long term follow up of all patients is required. Factors at initial evaluation indicative of risk of AC include a significant air-bone gap, otitis media with effusion in the contralateral ear and a smaller mastoid cavity ratio. The use of composite grafts at the time of CC surgery should be considered. Additionally, our findings suggest that the mastoid volume plays a causative role in the development of AC.

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ARTICLE INFO

Article history:
Received 7 July 2010
Received in revised form 23 September 2010
Accepted 1 October 2010
Available online 11 November 2010

ABSTRACT

Objectives: To determine the incidence of severe atelectatic otitis media and acquired cholesteatoma (AC) in children treated for congenital cholesteatoma (CC).

Methods: Retrospective chart review of 15 children who underwent primary surgery for CC over a 15 year period by a single surgeon.

Results: The mean postoperative follow up was 3.1 years. Significant tympanic retraction occurred in 6 children, included a retraction pocket that required T-tube insertion (3), and AC requiring tympanomastoid surgery (3). There was no complication related to retraction pocket in 9 children however 2 developed residual disease.

In comparing the two groups, those with and without subsequent significant tympanic retraction, both groups had similar gender, age, extent of CC (median Potsic grade of 2), bone erosion, and surgical technique. Differences were noted in air-bone gap at presentation (PTA 32.4 and 17.25), otitis media with effusion in the contralateral ear (3/6 and 1/9), smaller mastoid volume ratio compared with the contralateral ear (0.74 and 1.21), and longer average timing for second surgery (14.8 months and 8 months).

Conclusions: Acquired middle ear disease, including cholesteatoma, can follow surgical removal of CC, and long term follow up of all patients is required. Factors at initial evaluation indicative of risk of AC include a significant air-bone gap, otitis media with effusion in the contralateral ear and a smaller mastoid cavity ratio. The use of composite grafts at the time of CC surgery should be considered. Additionally, our findings suggest that the mastoid volume plays a causative role in the development of AC.

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identify predisposing factors and describing the treatment options applied to these challenging patients.

2. Materials and methods

A retrospective chart review was carried out at the Royal Children's Hospital, Melbourne, Australia, of children who underwent primary surgery for congenital cholesteatoma from January 1993 to December 2005. The follow up period incorporates until December 2006.

In order to include only patients that met strict diagnostic criteria, had a close and long term follow up and received a uniformed treatment, only private patients who were operated and followed up by the senior author (R.G.B.) were included. Fifteen cases were identified. The patients' medical records and CT scans were reviewed. Special attention was given to the type, details and outcomes of the initial surgery.

2.1. Primary assessment of congenital cholesteatoma

The criteria for diagnosis of congenital cholesteatoma, and inclusion in the study, were those described by Derlacki and Clemis [1], and modified by Levenson et al [6]: these were a pearly white mass, medial to an intact tympanic membrane; a normal pars tensa and flaccida; and no history of tympanic membrane perforation or previous otologic procedures. A history of prior bouts of otitis media did not exclude this diagnosis. In addition, every case included, had histological confirmation of cholesteatoma. Patients with previous history of otorrhea were excluded.

Extent of the disease at the time of surgery was quantified by means of which quadrants of the middle ear were involved and the degree of bone erosion found. In addition, the classification proposed by Potisic et al. was used [2]. This comprised: stage I with single quadrant involvement without ossicular involvement or mastoid extension; stage II with multiple quadrants involvement but without ossicular involvement or mastoid extension; stage III with ossicular involvement with erosion of ossicles requiring surgical removal for eradication of disease but without mastoid involvement; stage IV with disease extending to the mastoid.

2.2. Subsequent assessment of tympanic membrane retraction and acquired cholesteatoma

In order to exclude cases of recurrent or residual congenital cholesteatoma, subsequent development of acquired cholesteatoma were diagnosed using strict criteria. Only cases of primary acquired cholesteatoma were included, and those related to implantation in association with tympanostomy tube placement were excluded.

In summary all of the following criteria were required: (1) otoscopic or microscopic appearance of a retraction pocket, either in the pars tensa or pars flaccida, that accumulates keratin debris not manageable by suction, (2) complete removal of the congenital cholesteatoma during the primary surgery, (3) no anatomic relationship between the cholesteatoma and a preexisting tympanic membrane perforation or tympanostomy tube, and (4) histological confirmation of cholesteatoma after removal of the lesion.

The classification used to describe the degree of pars tensa retraction was the one proposed by Sade and Halevy [7] who described four stages of tympanic retraction: stage I, retracted membrane; stage II, retraction onto the incus; stage III, middle ear aterelactasis; stage IV, adhesive otitis media. A sub classification was also employed, based on the quadrant involved. Pars flaccida retraction was classified according to Sade et al. [8]. Grade 0, normal pars flaccida without retraction; Grade 1, slight retraction that does not touch the neck of the malleus; Grade 2, retraction lying against the neck of the malleus; Grade 3, partial destruction of the scutum; Grade 4, deep keratin-containing retraction pocket not manageable by suction, i.e., an attic cholesteatoma.

2.3. Assessment of mastoid pneumatization

The level of mastoid pneumatization was determined by reviewing available CT scans images of the patients that were obtained before their primary surgery. With no established criteria to grade mastoid pneumatization based on CT scans, the classification by Sade et al. [8], was adapted and simplified from plain radiographs, such as dividing patients in the following groups: Level I, sclerotic mastoid or diploic mastoid; Level II, small mastoid pneumatization, with several small air cells around the antrum; Level III, medium or large mastoid pneumatization.

Additionally, since most images were not transferable to a digital media, mastoid volumes were estimated using the formula for calculating a pyramid volume; this is using the largest measurements obtained in the 3 planes, axial, coronal and sagittal, and after summoned are divided in 3. A senior radiologist calculated all these measurements. By this, we could obtain the approximate volume of the mastoid air cell system, using the same procedure for all images. A mastoid volume ratio was further calculated by dividing the mastoid volume on the affected side with the mastoid volume obtained in the contralateral ear.

3. Results

Fifteen cases that met the inclusion criteria were identified. Two groups were clearly defined during the follow up period after CC removal.

Group 1 (6 children) consisted of patients that presented acquired middle ear pathology, such as atelectatic tympanic membranes, retraction pockets, or acquired cholesteatoma. Most of the patients in this group required multiple surgical procedures (median of 4).

Group 2 (9 cases) consisted of patients that had a predictable progress after the removal of the CC, they either, did not present any complication (7 patients), or it was purely related to the presence of residual disease (2 patients). Accordingly they required less surgical procedures (median of 1.5).

Although statistical analysis is limited by the size of the sample, both groups had similar composition in terms of gender (1 girl, 5 boys versus 2 girls, 7 boys), age (average 4.6 years versus 4.4 years), extension of initial disease (median Potsic grade 2 in both groups), bone erosion, and technique employed during primary surgery (atticotomy/mastoidectomy in 3 of 6 patients in group 1 versus 5 of 9 in group 2). Table 1 summarizes the main demographic and primary surgery findings and details of these groups.

Acquired middle ear complications in group 1 consisted of 3 patients (1, 3 and 4) that developed retraction pockets and were managed satisfactorily by 1 or more long lasting middle ear ventilation tubes insertion (T tubes), and 3 patients (2, 5 and 6) that required tympanomastoid surgery with removal and repair for deep retraction pockets and acquired cholesteatoma. Table 2 summarizes the main clinical features and management of these patients.

Between the groups, the following differences were noted: the air-bone gap was significantly higher in group 1 (PTA 32.4 versus 17.25), as well as the incidence of otitis media with effusion in the contralateral ear at presentation (3 out of 6 versus 1 out of 9) and the need of ventilation tubes insertion at that time (3 out of 6 versus 1 out of 9).

When comparing mastoid volumes, group 1 patients presented a smaller volume ratio than group 2 patients (0.74 versus 1.21).
Table 1
Demographic and primary surgery findings.

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Years of follow up</th>
<th>Gender</th>
<th>Age at diagnosis (years)</th>
<th>Symptoms at presentation (1)</th>
<th>Side involved</th>
<th>PTA preop (2)</th>
<th>Areas involved (3)</th>
<th>Potsic classification</th>
<th>OSSicular chain erosion (4)</th>
<th>Initial surgical procedure (5)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group 1</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>4.75</td>
<td>M</td>
<td>1.66</td>
<td>HL/OME</td>
<td>L</td>
<td>n/a</td>
<td>1, 3</td>
<td>2</td>
<td>0</td>
<td>ME + VT</td>
</tr>
<tr>
<td>2</td>
<td>5.66</td>
<td>M</td>
<td>5.66</td>
<td>HL/RAOM</td>
<td>R</td>
<td>25</td>
<td>1, 2, 3, 9</td>
<td>2</td>
<td>0</td>
<td>ME + VT</td>
</tr>
<tr>
<td>3</td>
<td>10.8</td>
<td>M</td>
<td>5.25</td>
<td>Unilat HL</td>
<td>R</td>
<td>35</td>
<td>2, 4, 9, 10, 11</td>
<td>3</td>
<td>1, 5</td>
<td>ME + CWU + r/o HM + MP</td>
</tr>
<tr>
<td>4</td>
<td>4.17</td>
<td>F</td>
<td>2.58</td>
<td>HL</td>
<td>L</td>
<td>30</td>
<td>1, 2, 9</td>
<td>2</td>
<td>0</td>
<td>ME + A + r/o M</td>
</tr>
<tr>
<td>5</td>
<td>5.66</td>
<td>M</td>
<td>7.83</td>
<td>Unilat HL</td>
<td>R</td>
<td>60</td>
<td>1, 2, 3, 4, 6, 8, 9, 10</td>
<td>3</td>
<td>1, 3, 4, 5</td>
<td>ME + CWU + r/o M + i + SS missing</td>
</tr>
<tr>
<td>6</td>
<td></td>
<td>M</td>
<td>4.66</td>
<td>Asymptomatic</td>
<td>R</td>
<td>12</td>
<td>1, 9</td>
<td>2</td>
<td>0</td>
<td>ME + MP</td>
</tr>
<tr>
<td>Partial results</td>
<td>5:1</td>
<td></td>
<td>Mean = 5.5</td>
<td></td>
<td></td>
<td>Mean = 4.6</td>
<td></td>
<td></td>
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<td>Group 2</td>
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<td></td>
<td></td>
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<tr>
<td>7</td>
<td>1.1</td>
<td>M</td>
<td>5.83</td>
<td>Asymptomatic</td>
<td>L</td>
<td>8.33</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>ME</td>
</tr>
<tr>
<td>8</td>
<td>0.66</td>
<td>F</td>
<td>5.83</td>
<td>Mass behind TM</td>
<td>R</td>
<td>D</td>
<td>1, 2, 3</td>
<td>2</td>
<td>0</td>
<td>ME</td>
</tr>
<tr>
<td>9</td>
<td>0.1</td>
<td>M</td>
<td>5.66</td>
<td>Unilat hearing loss</td>
<td>R</td>
<td>30</td>
<td>1, 2, 3, 4, 6, 9, 10, 11</td>
<td>4</td>
<td>1, 3, 4</td>
<td>ME + A + r/o M</td>
</tr>
<tr>
<td>10</td>
<td>1.75</td>
<td>F</td>
<td>3.92</td>
<td>RAOM</td>
<td>R</td>
<td>20</td>
<td>1, 2, 3, 4, 7, 9, 10</td>
<td>3</td>
<td>0</td>
<td>ME + MP</td>
</tr>
<tr>
<td>11</td>
<td>1</td>
<td>M</td>
<td>3</td>
<td>Asymptomatic</td>
<td>L</td>
<td>20</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>ME + MP</td>
</tr>
<tr>
<td>12</td>
<td>3.66</td>
<td>M</td>
<td>3.08</td>
<td>HL/OME</td>
<td>L</td>
<td>ID</td>
<td>1, 2, 3, 4, 9, 10, 11</td>
<td>4</td>
<td>0</td>
<td>ME + CWU</td>
</tr>
<tr>
<td>13</td>
<td>1.1</td>
<td>M</td>
<td>2.33</td>
<td>Poor speech/7HIL</td>
<td>L</td>
<td>20</td>
<td>1, 2, 3, 4, 9</td>
<td>2</td>
<td>0</td>
<td>ME + A + VT</td>
</tr>
<tr>
<td>14</td>
<td>3.42</td>
<td>M</td>
<td>3.92</td>
<td>Dizziness</td>
<td>R</td>
<td>15</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>ME</td>
</tr>
<tr>
<td>15</td>
<td>1</td>
<td>M</td>
<td>6.42</td>
<td>Unilat HL</td>
<td>R</td>
<td>32</td>
<td>1, 2, 3, 4, 9, 10</td>
<td>3</td>
<td>1,3,4</td>
<td>ME + A + r/o HM</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Mean = 7.2</td>
<td>Mean = 4.4</td>
<td></td>
<td>Mean = 2</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Mean = 3.1</td>
<td></td>
<td></td>
<td>Mean = 4.5</td>
<td></td>
<td></td>
<td>Mean = 5</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Mean = 4.4</td>
<td></td>
<td></td>
<td>Mean = 22.66</td>
<td></td>
<td></td>
<td>Mean = 2</td>
<td></td>
</tr>
</tbody>
</table>

(1) HL, hearing loss; OME, otitis media with effusion; RAOM, recurrent otitis media; unilat: unilateral; TM, tympanic membrane.

(2) n/a, not available.

(3) Areas included: 1, supero-anterior quadrant; 2, supero-posterior quadrant; 3, antero-inferior quadrant; 4, postero-inferior quadrant; 5, eustachian tube occlusion; 6, hypotympanum; 7, malleus–incudostapedial joint; 8, facial recess–sinus tympani; 9, anterior attic; 10, posterior attic; 11, aditus; 12, mastoid.

(4) I, long process of incus; 2, all incus; 3, capitulum of stapes; 4, crura of stapes; 5, head of malleus; 6, all malleus.

(5) ME, middle ear exploration and removal of CC; VT, ventilation tube; CWU, canal wall up mastoidectomy; r/o I, removal of incus; r/o HM, removal of head of malleus; r/o M, removal of malleus; i, incus; SS, stapes superstructure; MP, myringoplasty; A, atticotomy.
<table>
<thead>
<tr>
<th>Patient number</th>
<th>Contralateral middle ear pathology</th>
<th>Removal of CC</th>
<th>Early anatomical outcome</th>
<th>Timing between 1st and 2nd surgery (months)</th>
<th>Best PTA postoperative</th>
<th>2nd surgical procedure (1)</th>
<th>Number of total surgical procedures required</th>
<th>Acquired middle ear pathology</th>
<th>Summary of clinical progress (2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>OME</td>
<td>Complete removal</td>
<td>VT patent</td>
<td>16</td>
<td>22</td>
<td>VT</td>
<td>7</td>
<td>Atelectatic otitis media + contralateral cholesteatoma</td>
<td>L CC that presented bilateral AS retractions, which was stabilized by T-tube in the L, but developed a R aq cholest, later he developed COM, and finally received ossiculoplasty + MP</td>
</tr>
<tr>
<td>2</td>
<td>OME</td>
<td>Complete removal</td>
<td>TM intact, anterior retraction</td>
<td>19</td>
<td>ID</td>
<td>VT below retraction</td>
<td>4</td>
<td>Cholesteatoma</td>
<td>R CC that despite the use of VT progressed to a RP/cholesteatoma in the area of the previous CC and required TP + VT T-tubes to keep it stable</td>
</tr>
<tr>
<td>3</td>
<td>0</td>
<td>Complete removal</td>
<td>TM intact</td>
<td>16</td>
<td>25</td>
<td>VT</td>
<td>4</td>
<td>Atelectatic otitis media</td>
<td>R extensive CC, later chronic retraction of TM, required VT2 times</td>
</tr>
<tr>
<td>4</td>
<td>OME</td>
<td>? Residual disease</td>
<td>TM intact</td>
<td>8</td>
<td>23</td>
<td>ME + MP</td>
<td>4</td>
<td>Atelectatic otitis media</td>
<td>LCC, that had small residual disease (pearl), removed at 2nd look surg, later presented chronic OME in the R – 3 sets of VT and RP in the Lthat req T-tubes</td>
</tr>
<tr>
<td>5</td>
<td>0</td>
<td>Complete removal</td>
<td>TM intact</td>
<td>13</td>
<td>52</td>
<td>ME + TORP</td>
<td>3</td>
<td>Cholesteatoma</td>
<td>R extensive CC required CWU/2nd look = no residual disease – TORP. 4years later presented postsuperior RP and attico-mastoid cholesteatoma required another CWU</td>
</tr>
<tr>
<td>6</td>
<td>0</td>
<td>Complete removal</td>
<td>TM intact</td>
<td>17</td>
<td>23</td>
<td>ME + MP (composite graft)</td>
<td>2</td>
<td>Cholesteatoma</td>
<td>RCC that years later presented with a severe RP/cholesteatoma, needed removal of these pockets</td>
</tr>
</tbody>
</table>

Total

Mean = 14.8
Mean = 25.8

(1) TORP, total ossicular replacement prosthesis; ME, middle ear exploration and removal of disease; MP, myringoplasty; composite graft = cartilage and pericondrium.
(2) L, left; R, right; AS, antero-superior; COM, chronic otitis media; TP, tympanoplasty.
requiring recurrent tympanostomy tubes would have developed a
and safe ears. In fact we can hypothesize that some of the patients
correctly and uniformly managed, they will end with good results
procedures, if the patients continue with a long follow up, and are
this review shows that even with complications and multiple
long term complications, such as acquired cholesteatoma, which
patients is required, since congenital cholesteatoma is a challeng-
allowing the disease to present in a more advance stage.

Our data supports and expands a report by Lazard et al., in
that acquired middle ear disease can follow surgical removal of CC,
but differs in the following issues: (1) for the first time we present
cases of severe RP and acquired cholesteatoma, that required
multiple procedures to achieve a safe ear and/or serviceable
hearing, (2) the relative frequency of acquired pathology in CC
patients in our series is 40%, much bigger than the 13.7% presented
by them, (3) in our series contralateral ear effusion was related
with acquired pathology, no such relation was found by them, (4)
their report included 116 patients, much more than our report of
15, (5) our data represent the uniformity of one single profession-
als criteria as opposed to the diversity of criteria of a whole
department, and (6) they report that Eustachian tube extension
and lack of cartilage reconstruction of attic defects were related to
later acquired pathology, but no such relation was seen in our
patients.

The similitudes and differences between both reports empha-
size the importance of awareness about these complications,
and the need of further studies to address this problem.

Two patients developed residual disease, they followed the
typical course of such patients, namely: (1) their disease was more
advanced, as clearly shown by large “Potsic grades” of 3 and 4,
compared with the median of 2 in the rest of the patients, (2) there
was the opinion at the moment of primary surgery that a complete
removal was not achieved and (3) the unsatisfactory results were
evident soon after the primary operation, and needed to receive a
secondary procedures 2 and 7 months later. These characteristics
remarkably contrast with the clinical course patients in group 1
experienced, they had less extensive disease (median Potsic grade
of 2), in all cases a complete removal was achieved in the first
attempt (Table 2) and with the exception of 1 case, all presented a
late complication and the secondary procedure was performed
more than a year after the primary surgery. All these observations
reassure us that patients in group 1 truly experienced the outbreak
of new pathology, i.e. acquired RP and/or cholesteatoma and do not
represent residual or recurrent disease.

As stated previously, there are patient-related features that
correlate with acquired middle ear disease in this group of
patients. Although Tympanometry was not uniformly obtained in
this series, it would have been useful to indicate the degree to
which tympanic membrane retraction was due to impaired
Eustachian tube function or atrophy of the tympanic membrane.

Nevertheless, iatrogenic factors should also be considered as
inductors of such progression.

Possible factors include, but are not limited to: (1) disruption of
the blood supply to the antero-inferior quadrants of the tympanic
membrane by means of the placement of the tympanomeatal flap
incision, (2) structural damage to the tympanic membrane by
means of totally or partially separating the tympanic membrane
from the malleus, (3) middle ear or attic aeration disfunction
created by post-surgical adhesions at the level of the Eustachian
tube or Pruskas’s space, (4) lack of stability by partial or total loss of
ossicular chain or inadequate posterior canal wall reconstruction,
and (5) grafting material used. Lazard et al. [5] reported that
Eustachian tube extension of CC and failure to properly reconstruct
an atticotomy relate to the appearance of RP. But they also admit
that RP occurred despite cartilage reinforcement of the tympanic

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Table 3 summarizes the mastoid measurements obtained from CT
scan data and their ratio.

Lastly, group 1 patients had a longer period of time between
first and second surgery (average of 14.8 months versus 8 months),
indicating that group 1 patients consisted mainly of patients that
had complete resolution of their CC, and only developed acquired
disease later in their course.

4. Discussion

15 patients collected during a 15 years period may seem a small
group, specially when compared with most published series,
which with some exceptions [5], usually present results of about
50 patients [9–11]. However, in an effort to be specially strict with
diagnostic, therapeutic and evaluation criteria presented, we
decided to include only private patients who were operated and
followed up by the senior author (R.G.B.) and not all cases seen at
our institution. On the sacrifice of number of patients we opted for
reliability. This is particularly important in our case, where we are
reporting acquired complications in CC patients, and the doubt of
veracity of the initial diagnosis is always present [5]. For this same
reason we excluded patients that presented with initial otorrhea,
and the diagnostic criteria suggested by Derlacki and Clemis [1]
and Levenson et al. [6] were strictly followed.

Moreover, this series, by been seen by a single practitioner,
represent a unique group in CC reports, and since 3 patients
presented to the outpatients clinic with no related symptoms, it
highlights the importance of a thoughtful examination on every
single patient. Our data suggest that a meticulous paediatric
otolaryngologist should find one congenital cholesteatoma per
year, and if he or she does not find them is probably missing them,
allowing the disease to present in a more advance stage.

Our data supports the fact that a long term follow up of all
patients is required, since congenital cholesteatoma is a challeng-
ing condition, very diverse and that can even be associated with
long term complications, such as acquired cholesteatoma, which
demands the best quality of care from our team. On the other hand,
this review shows that even with complications and multiple
procedures, if the patients continue with a long follow up, and are
correctly and uniformly managed, they will end with good results
and safe ears. In fact we can hypothesize that some of the patients
requiring recurrent tympanostomy tubes would have developed a
cholesteatoma if not being carefully followed up. Since in our
experience more than one third of patients operated for CC will
develop a significant middle ear disease later, such a long and close
follow up of all patients is guaranteed, nevertheless special
attention should be given to patients that at the moment of the
primary surgery and regardless of the surgical technique
employed, present with a significant air-bone gap, otitis media
with effusion in the contralateral ear and a smaller mastoid cavity,
compared with the contralateral one.

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Mastoid volume</th>
<th>Mastoid volume ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>affected ear (cm³)</td>
<td>contralateral ear (cm³)</td>
</tr>
<tr>
<td>Group 1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>4.5</td>
<td>2.4</td>
</tr>
<tr>
<td>2</td>
<td>n/a</td>
<td>n/a</td>
</tr>
<tr>
<td>3</td>
<td>4.4</td>
<td>4.6</td>
</tr>
<tr>
<td>4</td>
<td>0.5</td>
<td>0.8</td>
</tr>
<tr>
<td>5</td>
<td>0.1</td>
<td>4.8</td>
</tr>
<tr>
<td>6</td>
<td>2.6</td>
<td>12.9</td>
</tr>
<tr>
<td>Partial results</td>
<td></td>
<td>Mean = 0.74</td>
</tr>
<tr>
<td>Group 2</td>
<td></td>
<td></td>
</tr>
<tr>
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<td>n/a</td>
<td>n/a</td>
</tr>
<tr>
<td>8</td>
<td>n/a</td>
<td>n/a</td>
</tr>
<tr>
<td>9</td>
<td>n/a</td>
<td>n/a</td>
</tr>
<tr>
<td>10</td>
<td>3.7</td>
<td>7.8</td>
</tr>
<tr>
<td>11</td>
<td>4.4</td>
<td>4.1</td>
</tr>
<tr>
<td>12</td>
<td>0.7</td>
<td>2.4</td>
</tr>
<tr>
<td>13</td>
<td>1</td>
<td>0.6</td>
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<tr>
<td>14</td>
<td>8.28</td>
<td>3.1</td>
</tr>
<tr>
<td>15</td>
<td>5.3</td>
<td>4.8</td>
</tr>
<tr>
<td>Partial results</td>
<td></td>
<td>mean = 1.21</td>
</tr>
</tbody>
</table>

n/a, not available.
membrane in 56.25% of cases and that was impossible to differentiate iatrogenic from patient induced factors. We could not find any surgical-induced factor particularly in group 1, and feel as they did, that “one could propose the systematic reconstruction of any destroyed part of the canal wall (spontaneous or iatrogenic) during surgery for CC, with particular attention being paid to its posterior and superior parts” [5].

At the same time, in light of our findings we should suggest that patients presenting with significant air-bone gap, otitis media with effusion in the contralateral ear and a smaller mastoid cavity ratio should be considered for prophylactic tympanostomy tube insertion at the same time of primary surgery or receive a composite graft with cartilage reinforcement. In the later case since microscopic examination will be limited in the follow up period, MRI scan [12] or routine second look surgery should be taken into account as well.

Additionally, our findings represent a unique collection of CT scans collected several months before the appearance of acquired cholesteatoma, and therefore enabled us to examine, with prospectively obtained information, the hypothesis first suggested by Sade et al. [8] about the deficiency of the mastoid air cell system pressure buffer for acquired cholesteatoma.

The hypothesis theorizes that Eustachian tube dysfunction alone is not the responsible for the development of retraction pocket cholesteatomas, but rather Eustachian tube dysfunction, together with a non well developed mastoid cavity create the chronic conditions of negative pressure that will result in a RP cholesteatoma.

It is noteworthy that patient 1 presented a smaller mastoid volume on the contralateral side of the congenital cholesteatoma, and 8 years later presented an acquired cholesteatoma on that side.

Therefore if we would like to analyze the mastoid volume as a risk factor for future development of retraction pocket cholesteatomas we should add this case to the other 3 patients that developed an acquired cholesteatoma on the congenital cholesteatoma side, by inverting his mastoid ratio. In that case this results in a ratio of 0.53, and together with the available data of patients 5 and 6 (0.02 and 0.2 respectively) they represent the smallest ratios in group 1, and consequently supporting the theory of the mastoid air cell system as a pressure buffer. However, with the limited number of CT scans available it is impossible to draw definitive conclusions, specially when noticing that patients 10 and 12 in group 2 also presented small ratios, and more studies on this direction should be performed.

References